Case Report

Askin Tumour: A Rare Thoracopulmonary Tumour in Adults

Vikas Sikri and Sanjay Sobti

Department of Respiratory Medicine, Indraprastha Apollo Hospital, New Delhi, India

ABSTRACT

Askin tumour, a primitive neuroectodermal tumour of the thoracopulmonary region, is a rare tumour presenting in childhood. Its presentation in adults is rare. We report a case of an Askin tumour in an adult patient who presented to us with worsening breathlessness and vague chest pain. Investigations including immunohistochemistry confirmed the diagnosis of Askin tumour. **[Indian J Chest Dis Allied Sci 2013;55:233-235]**

Key words: Askin, Primitive neuroectodermal tumour, Thoracopulmonary.

INTRODUCTION

Askin tumour was defined by Askin and Rosai[1] in 1979. Although once accepted as distinct entity, Ewing's sarcoma, Askin tumour and primitive neuroectodermal tumour (PNET) are all considered as members of the Ewing's family of tumours; and when localised to the thoraco-pulmonary region, these are termed as Askin tumours.[1] The frequency of Ewing's sarcoma and PNET among childhood tumours is 2%, the frequency of occurrence of Askin tumour is established due to the rarity of the disease. Askin tumour is a rare tumour of childhood and usually presents with common respiratory symptoms. Its occurrence in adults is rare. However, we are reporting here a case of an adult patient with Askin tumour.

CASE REPORT

A 46-year-old male school teacher, resident of Sirsa (Haryana), smoker, non-alcoholic, presented with cough and nasal symptoms for the past three months. He was treated symptomatically with oral antibiotics and was partially relieved. However, a week later he developed progressive breathlessness and vague chest pain over the left subcostal region.

Chest radiograph (posterior-anterior view; Figure 1) revealed a left paratracheal lesion. Computed tomography of the chest (Figure 2) showed a middle mediastinal mass lesion adjacent to the left pulmonary artery that was not infiltrating the lung. The patient was referred to our hospital for further management.

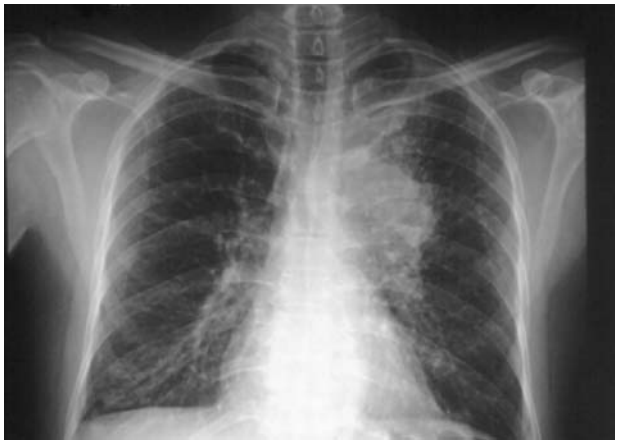

Figure 1. Chest radiograph (postero-anterior view) showing a left paratracheal mass.

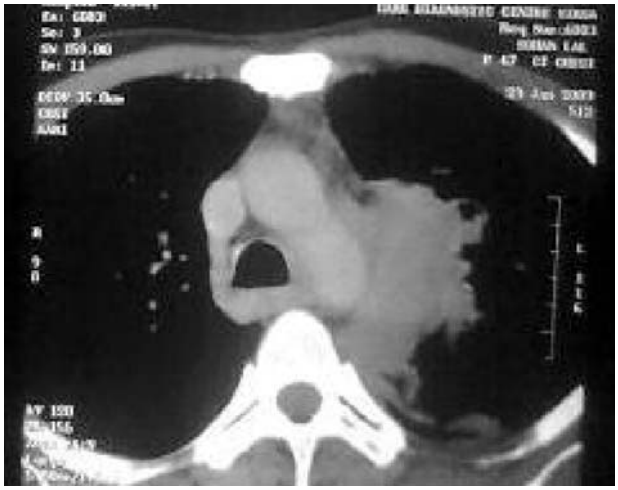

Figure 2. Computed tomography of the chest (mediastinal window) showing a tumour mass adjacent to the left pulmonary artery.

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Correspondence and reprint requests: Dr Vikas Sikri, 161, LIG Flats, Sarita Vihar, Mathura Road, New Delhi-110 076, India; Phone: 09818900866; Fax: 91-11-26823629; E-mail: vschestdoc@gmail.com

On examination, his vital parameters were stable. Neck examination revealed a palpable left supra-clavicular lymph node that was discrete and hard on palpation. Bilateral wheeze and left basal crackles were noted on auscultation of the chest. Haematology and blood chemistry were within normal limits. He was started on empirical antibiotics, bronchodilators and analgesics.

Fine needle aspiration cytology (FNAC) of the lymph node revealed it to be a small cell carcinoma.

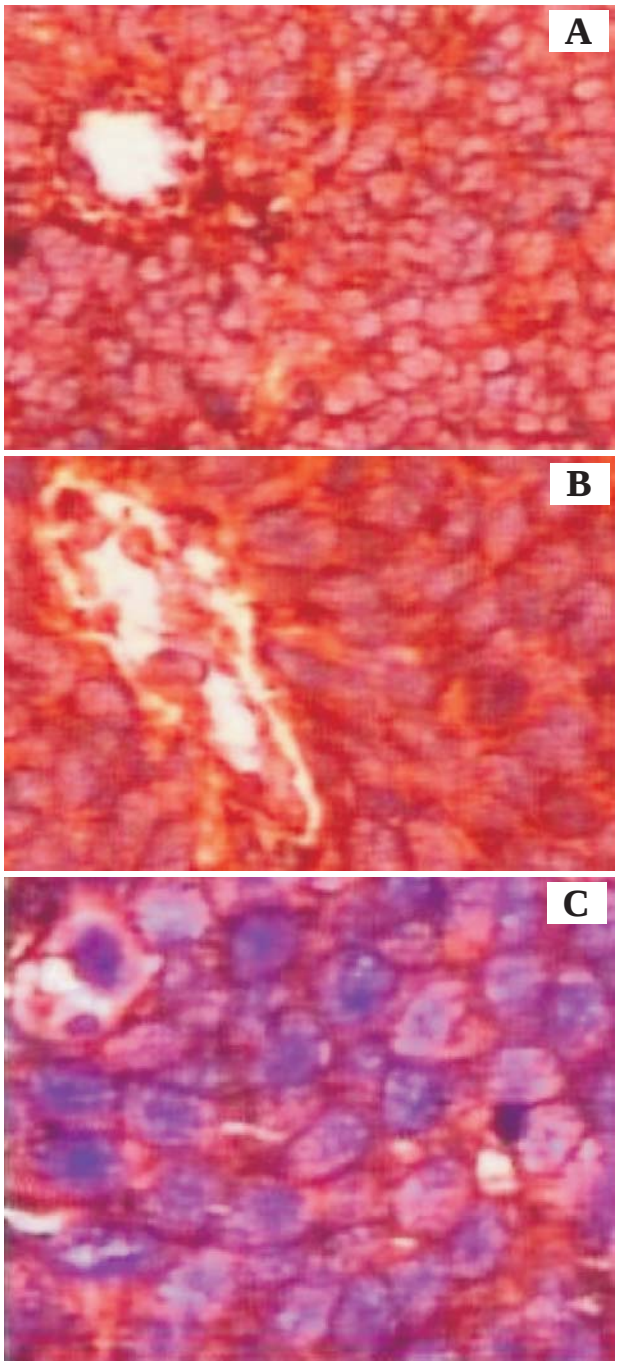

Figure 3. Immunohistochemistry photograph showing (A) tumour cells positive for synaptophysim(), (B) positive for vimentin(), and (C) positive for MIC-2.

Excision biopsy revealed almost complete replacement by a tumour composed of sheets of cells with scanty cytoplasm. The nuclei were hyperchromatic with "salt and pepper" chromatin and inconspicuous nucleoli. Nuclear moulding and brisk mitosis was noted. Perivascular pseudo-rosettes was seen at places.

Immunohistochemistry showed tumour cells positive for synaptophysin (Figure 3A), vimentin (Figure 3B) and MIC-2 (Figure 3C), while being negative for cytokeratin, LCA, S-100, TTF and Desmin.

The histology was consistent with a malignant round cell tumour. The overall morphology and immune profile of the tumour was consistent with metastasis from PNET. Contrast enhanced CT of the whole abdomen, brain and bone scan did not show any evidence of metastatic involvement. The patient was given six cycles of chemotherapy (vincristine, actinomycin D and cyclophosphamide) and concurrent radiotherapy. Three months after treatment, he had a relapse and chemotherapy was re-started. CT of the chest (Figure 4) revealed extensive pleural metastasis with satellite lesions in the right lung. The patient's condition deteriorated and he succumbed to his illness.

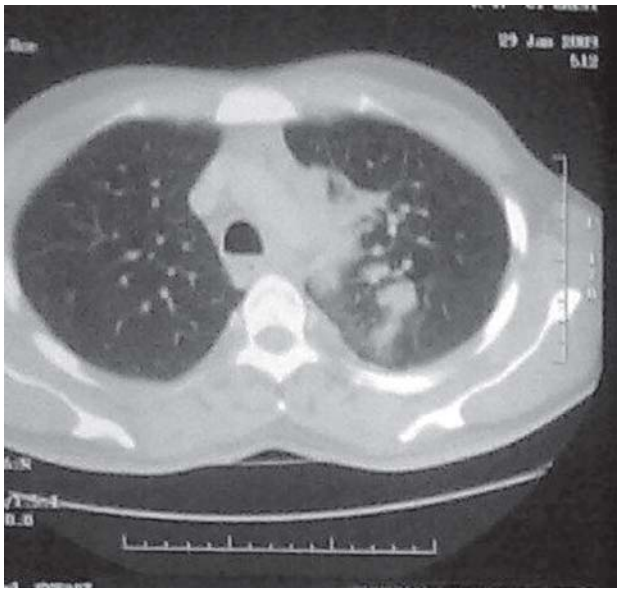

Figure 4. Computed tomography of the chest showing pleural metastasis with satellite mass lesion in the right lung.

DISCUSSION

Dickinson *et al*[2] in a 45-year database review found the prevalence of Askin's tumour to be of 0.2 cases per million. Ewing's sarcoma, PNET, rhabdomyosarcoma, neuroblastoma and lymphoma are small round cell tumours encountered in children and young adults.[3,4] Although Ewing's sarcoma and PNET can be easily differentiated microscopically from other entities in this group, the differentiation amongst these two is

difficult. This malignant round cell tumour, that originates from the soft tissue of the chest wall, is also called extra-skeletal Ewing's sarcoma or peripheral PNET. [5,6]

Typically, Askin tumour develops as a solitary mass, rarely involving most of the hemithorax[7] or as multiple masses in the thoraco-pulmonary region (thoracic wall, lung, mediastinum, or pericardium). Askin tumour, also named as 'extra-skeletal' Ewing's sarcoma' or 'soft tissue Ewing's sarcoma' arising from the soft tissue of the chest wall, shows a neural differentiation that can be demonstrated by immunohistochemical and ultrastructural methods. Similar to Ewing's sarcoma and PNET, these tumours have positivity for neural markers, such as neuron specific enolase and also neuroendocrine markers, such as chromagranin and synaptophysin. These are also positive for MIC-2 gene which produces CD 99 and a cell membrane-like protein p 30/32 which are highly sensitive but not specific products. According to the current consensus, the so-called Askin tumour is a variant of Ewing's sarcoma and PNET that involves the thoracopulmonary region.

Rarely, Askin tumours are found in the central nervous system. In the thoracic area, these tumours are invasive and prone to involve bone (ribs and scapula), invading the retroperitoneal space, and spreading to lymph nodes, adrenals, and liver. Askin *et a1*[1] reported that small round cell tumours of childhood and adolescence located in the thoraco-pulmonary region are more common in females, the median age for these being 14.5 years. Pain is the only or the main symptom in 60% of the cases. Radiological characteristics range from a unilateral chest wall mass to pleural fluid, invasion to the adjacent lung parenchyma, pulmonary nodules and sometimes lymphadenopathy. The diagnosis of Askin tumour rests on cytopathological investigations and immunohistochemical tests.

Treatment in Askin tumour consists of radical surgery, neo-adjuvant or adjuvant chemotherapy and radiotherapy. Although a long survival is intended by multimodel therapy, prognosis is generally poor. The best prognosis can be provided by surgical treatment with wide resection. Recurrences in the primary tumour site are important in differentiating these tumours from other tumours in children and adolescents.[1] As local recurrences after resection and metastases are frequently seen in Askin tumour, it has a poor prognosis and a short survival.[5] The most common recurrence sites are the skeleton, sympathetic chain and the original site. Indicators of poor prognosis include advanced age, metastatic disease, extraosseous primary tumour and reccurrence.[8]

Recent studies have shown that remission rate has improved from 26% to 65% with aggressive chemotherapy. Various chemotherapy regimens[9] have been used that include VAC (vincristine, actinomycin D, cyclophosphamide), VACA (vincristine, actinomycin D, cyclophosphamide, adriamycin) and VAC alternating IE (ifosamide and etoposide). Average survival has been reported to be eight months after the diagnosis.[1] As our case was inoperable in view of its middle mediastinal location, chemotherapy was given.

To conclude, Askin tumour should be considered as an aetiologic possibility in a small-cell tumour of the thoracopulmonary region, especially in the young age group. Patients with such tumours should be treated surgically, with wide local excision wherever possible. Combination chemotherapy should be considered in patients with inoperable disease.

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